Diencephalic cysts

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Four cases of extensive cystic dilatation of the third ventricle associated with agenesis of the corpus callosum are described. This condition is termed "diencephalic cyst"; the clinical and radiological features are described. The embryological development of the diencephalon, the telencephalon, and the corpus callosum are described in relation to this malformation. The associated hydrocephalus is also discussed. The treatment and clinical course of the patients is outlined.

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agenesis of corpus callosum hydrocephalus embryology

Congenital cysts of the diencephalon are among the rarer developmental abnormalities of the central nervous system, and have received little attention. This paper presents four examples of the condition, and suggests that it constitutes a clinical entity. The embryological development of the malformation probably lies in between the group of malformations associated with anterior neuropore closure and prosencephalization, and the group associated with occipital encephaloceles and abnormalities of the rhomboencephalon.

Case Reports

Case 1

This boy was first seen at the University of Kentucky Medical Center when he was 5 days old because he had a midline swelling situated over the posterior part of the sagittal suture. Over a period of 2 months the skull circumference became abnormally large, and the anterior fontanel and the cystic mass both became tense. The basic neurological examination was normal and the child robust. Skull films showed enlargement of the cranial vault and hypertelorism. A lumbar pneumoencephalogram demonstrated not only the typical ventricular pattern of agenesis of the corpus callosum, but also a large midline cyst which was in continuity with the third ventricle and extended upward between the hemispheres to reach the vault at the site of the cutaneous lesion (Fig. 1). An angiogram confirmed the features of agenesis of the corpus callosum, and showed that the pericallosal arteries were separated by the midline cyst. The venous angiogram demonstrated that the sagittal sinus passed backward on both sides of the interhemispheric cyst and was duplicated as far as the confluence of the sinuses, where there were a number of abnormal branches. At operation the scalp lesion was excised and its stalk followed in through the sagittal suture and between the duplicated limb of the sagittal sinus. The scalp cyst was demonstrated to be in continuity with the midline interhemispheric cyst, and a shunt was performed directly from the cyst to the right atrium. The histological examination of the scalp cyst showed a fibrous lining and no ependyma. The subsequent course of the child has been entirely satisfactory; at the age of 2½ years he was able to talk well and walk normally with good coordination. The sunken anterior fontanel was still open; skull circumference was 52 cm.

Case 2

This boy was seen when 4 months old by Dr. Robert McLaurin of the University of
Cincinnati who kindly permitted the inclusion of the patient in this report. He had a small midline lump in the interparietal region which had been present since birth, a somewhat enlarged skull, and a bulging anterior fontanel. The air studies at that stage demonstrated a midline cyst between the cerebral hemispheres, agenesis of the corpus callosum, and no dilatation of the ventricular system. Histological examination of the excised cyst showed fibrous tissue around an epithelial-lined cavity. At 1 year of age, the child was mentally retarded and the anterior fontanel still open. A further pneumoencephalogram demonstrated more clearly the dorsally placed midline cyst extending up into the interparietal region between the hemispheres and just beneath the sagittal suture. When the child was seen 8 years later at the University of Kentucky he was mentally retarded and had a large head.

Case 3
This girl was admitted to the University of Kentucky when 4 days old because of an enlarged head. She showed normal neurological function, skull films confirmed the enlargement of the calvarium plus flattening of the squamous occiput and an apparently shallow posterior fossa. A lumbar air encephalogram filled the basal cisterns and also a large posterior fossa cyst that extended upward toward the lambda to the level of the tentorial hiatus. A ventriculogram showed a
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hugely dilated third ventricle and the appearance of agenesis of the corpus callosum, and a diencephalic cyst which reached backward and upward to the sagittal area in the posterior interparietal region. The ventricles communicated with the posterior fossa cyst. An angiogram showed the confluence of the sinuses to be in an abnormally high position between the diencephalic cyst and the posterior fossa cyst. There was a common anterior cerebral artery filling from the right side and passing backward to divide around the diencephalic cyst; the venous sagittal sinus passed to the left of the cyst. A ventriculoperitoneal shunt was performed and subsequently revised to a ventriculoatrial shunt after a period of infection. Continued skull enlargement was shown by ventriculography to be due to persistent dilatation of the huge midline diencephalic cyst which was no longer in continuity with the lateral ventricles. The cyst itself was added to the ventriculoatrial shunt. Subsequent progress has been satisfactory; at the age of 22 months she was able to walk with an unsteady gait, could feed herself, and had spontaneous conversation of a few words strung together. Poor head control, an alternating internal strabismus, and slightly increased tone in the arms and legs has persisted.

Case 4

This girl was admitted to the University of Kentucky Medical Center at the age of 2 days because of a large head. A ventriculogram demonstrated that the lateral ventricles were dilated and separated, and the third ventricle continuous with a diencephalic cyst which reached the top of the cranial vault between the hemispheres (Fig. 2); air entered the dilated cisterna magna via a large fourth ventricle. An angiogram confirmed...
agenesis of the corpus callosum and demonstrated that the pericallosal arteries were separated by the diencephalic cyst (Fig. 2, lower left). In an isotope ventriculogram using radioactive-iodinated human serum albumin (RIHSA),\(^2\) the isotope remained in the ventricles. A ventriculostriatal shunt was performed, and subsequent progress has been satisfactory at short-term follow-up.

**Discussion**

These four patients all had in common a diencephalic cyst extending from the third ventricle to the sagittal region of the skull. In the first two cases, the diencephalic cyst was also associated with a midline cyst under the scalp, which in Case 1 was clearly demonstrated to be in continuity with the diencephalic cyst. The histology of the two scalp cysts did not indicate clearly that they arose from the ependyma of the third ventricle, and they could well have been fibrosed meningoceles.

The last two cases were associated with posterior fossa cysts of the variety described by Robertson\(^8\) as extensions of the cisterna magna and in Case 3 there appeared to be continuity between the posterior fossa and diencephalic cysts.

Studies of the development of the human embryo have shown that by the age of 30 days (the 4 mm, late 28-somite stage) the ventricular system has already developed to form the prosencephalon, mesencephalon, and rhomboencephalon.\(^1,5,4\) At the 10 mm (37-day) stage, the prosencephalon develops the telencephalic dilatation in a dorsolateral direction to form the future cerebral hemispheres, and the ventral median portion remains as the diencephalon. The future third ventricle is formed from the diencephalon and the portion of the telencephalon between the two intraventricular foramina known as the telencephalic medianum. In the 24 mm (7\(\frac{1}{2}\) weeks) embryo, the diencephalon is still a relatively large cavity and separated from the ectoderm by a thin layer of mesoderm. The side walls of the diencephalon become limited by the development of the thalamus, hypothalamus, and epithalamus, and the blending of these with the corpus striatum of the telencephalon as it develops dorsocaudally.

The dorsal limitation of the diencephalon is determined by the development of the commissures. The anterior and hippocampal commissures appear first, and are present at the 25 mm stage; they lie initially in a cephalic position continuous with the lamina terminalis. The corpus callosum appears much later at the 60 mm stage, and then develops slowly in a caudal direction to be completed by the 200 mm stage. In this way the corpus callosum comes to overlie the roof of the third ventricle and to form the dorsal boundary of its telachoroidea.

The diencephalic cyst is probably a mal-development established prior to the 60 mm stage, and constitutes a failure of the diencephalon to sink ventrally after anterior neuropore closure. The interparietal scalp cysts are really diencephalic encephaloceles. Agenesis of the corpus callosum would be the natural corollary of this development; this view is compatible with the recent extensive studies by Loeser and Alvord\(^5,6\) of the embryology and maldevelopment of this structure. It seems reasonable to classify these diencephalic cyst malformations as lying between the group associated with anterior neuropore closure and prosencephalization (anterior encephaloceles and holoprosencephaly) and the group associated with maldevelopment of the hindbrain (cerebellar and fourth ventricle encephaloceles and Dandy Walker cysts of the rhomboencephalon).

The mechanism for the development of hydrocephalus in our cases is difficult to understand; the radiological studies of these patients indicated a communicating type of hydrocephalus, and the one dynamic study using RIHSA ventriculography showed that the albumin did not leave the lateral ventricles. It is possible that the intracranial diencephalic cysts are in themselves obstructing lesions because of their cul-de-sac nature, as has been suggested in the case of arachnoidal cysts.\(^7\) Another possibility is that the choroid plexus at the base of the diencephalic cyst continues to secrete the fluid, which expands the cyst, as apparently occurred in Case 3 when the cyst became separated from the ventricular system.

The limited observations on the progress of these four children suggest that the prognosis is not necessarily poor, and that the scalp cyst should be excised and the associated hydrocephalus treated.
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Summary

Four examples of "diencephalic cyst" have been presented as a possible distinct entity, with or without communication with another cyst under the scalp, or in the posterior fossa. Hydrocephalus was associated with the condition, and when treated, the subsequent neurological progress of the child was often promising.

References


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